

A 42-YEAR-OLD KING WITH A CAVITARY PULMONARY LESION AND INTESTINAL PERFORATION

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INTESTINAL tuberculosis, usually involving the terminal ileum and cecum, may occur as a primary manifestation or as a complication of primary pulmonary tuberculosis. One of the aphorisms of Hippocrates warned that "phthisical persons die if diarrhea sets in." Prior to the introduction of chemotherapy, intestinal ulceration with fistula formation was a common finding at autopsy in patients with active cavitory lesions;¹ tuberculous peritonitis was a rare complication.² The association of intestinal ulceration, free perforation, and peritonitis with a cavitory pulmonary lesion was vividly described in 1643 in the autopsy report of Louis XIII.³

We have found the omentum eroded, the small intestines enormously swollen, diminished in color and bathed in a quantity of muddy, purulent, serous fluid, the external surface of the liver pale, as if it had been boiled, the stomach filled with a blackish fluid that stained its lining and containing one worm approximately 15 cm. in length and several other small worms, the duodenum huge and filled with bilious material, the jejunum is filled with the same material and entirely yellow inside, the ileum less tinted and less filled with a thicker material, the cecum from its origin is red and devoid of its meaty membrane, the process continuing with increasing severity until the end of the colon where an ulcer that pierced the intestine is caused by the descent of the mud exuding from the inferior mesentery that is ulcerated in several places from which more than a jug-full of spilled, purulent material accumulates throughout the abdomen, bathing the intestines. In addition to the color of the liver described above we found that its concave surface, which tears when touched, to be deprived of its capsule, upon being cut it appears dried and recooked on the inside as on the outside. In the right kidney a small abscess filled with green mud is enclosed in a cyst in its inferior meaty portion. All of the left lung, and less of the right, is entirely attached to the ribs. The superior portion of the left lung contains a large ulcerated cavity filled with mud. All of these events were recognized as true causes of his death.

Performed at Saint-Germain at 6 o'clock in the morning, May 15, 1643.

His attending physicians displayed an interest in the postmortem that is regrettably absent among many of our contemporaries. Present at the autopsy were Charles de Savoye, Nicolas of The Vitry Hospital, de Souvré, Bouvart (first physician to the King, who dictated the autopsy report), Seguin (first physician to the Queen Regent), Vaultier (first physician to the deceased Queen Mother), Chicot and Conrade (physicians to the King when he travelled), de la Vigne (regent and dean of the Faculty of Medicine of Paris), Moreau (a physician of the faculty who recorded the protocol), Yvelin (physician to the regent queen), Brunyer and de Nugent (physicians to the duke of Orleans), Baptiste Bontemps (first surgeon to the king), Pescheval (first surgeon to the Queen), Collart (first surgeon to the duke of Orleans), Regnault, Lycot and Alexander le Roy (surgeons serving the king who performed the autopsy), Colin and le Large, surgeons who assisted at the autopsy. All but Brunyer and le Large signed the protocol.

Although this is considered one of the earliest reports of secondary intestinal tuberculosis, it is difficult to reconcile Louis XIII's extensive clinical record and the postmortem findings in the bowel with this diagnosis. For 16 years, from the age of 26 until his death, Louis XIII suffered from repeated attacks of febrile gastroenteritis, diarrhea with bloody stools containing pus and mucus and arthritis. His disease began in 1627 with several episodes of abdominal distention, fever, and dysentery. Arthritic manifestations appeared the following year. In 1629 he was ill again with diarrhea and arthritis. Louis XIII almost died in 1630 and in one 24-hour period he was reported to have had 40 bowel movements, many of them containing pure blood, "as if from a vein" and a rectal abscess. His physicians despaired, but, "as if inspired by God," they decided to perform a final (the seventh) phlebotomy directly from his right arm. This apparently was effective; his diarrhea subsided and the king recovered. Loss of appetite, vomiting, and insomnia occurred again in 1631. A cough was described for the first time in 1632; however, his intestinal complaints and arthritis predominated. After another such episode in 1634, Louis XIII had three years of good health, and then diarrhea and rheumatism of the right knee recurred. Louis XIII's tongue was described by his valet de chambre as "a tongue so long and so thick that when it was out of his mouth he had difficulty in returning it and was obliged to push it in with his finger." His attacks of intestinal disease became more frequent and in 1642 he had several episodes of severe abdominal pain,

fever, diarrhea with a bloody membrane in his stools, and something in the anus which may have been an "abscess or hemorrhoids."

Although it is not unlikely that Louis XIII had pulmonary tuberculosis, neither the prolonged course of active bowel disease nor the appearance of the bowel at autopsy are consistent with intestinal tuberculosis. In a recent study of 13 cases of intestinal tuberculosis, the gross morphologic findings in all were of a mass in the right lower quadrant made up of inflamed, thickened cecum with adherent loops of bowel.⁴ When perforation occurs it is almost always in the cecum. The thinned bowel wall and perforation in the colon found in Louis XIII are not typical of tuberculous enteritis. Other possibilities which have not been considered include ulcerative colitis and chronic amebic colitis. Although the latter entity cannot be excluded, chronic amebic colitis is associated with fibrosis that may be so severe that it results in stricture and obstruction. It is difficult to interpret the description of the liver, but nothing is described that resembles an amebic abscess. The location of the cavitary lesion in the left upper lung also does not favor an amebic infection. Louis XIII's autopsy findings and clinical history make it more likely that he suffered from ulcerative colitis. The king's macroglossia most probably reflects an infiltration of amyloid secondary to colitis or pulmonary tuberculosis.

The concept that ulcerative colitis results from emotional trauma is unproved. Nevertheless, bereavement and early separation from a person on whom the patient depends, sensitivity to humiliation, parental disapproval, and a sense of helplessness or inability to cope have been described as characteristics of patients with ulcerative colitis.⁵ All of these apply to Louis XIII. Louis XIII's early life and medical history are recorded in extraordinary detail. Born in 1601, he was the first legitimate son of Henry IV and the first child born a dauphin of France in 84 years. After proudly displaying the infant to the hundreds of courtiers assembled outside of the Queen's chamber, the king immediately delivered him to the care of Dr. Jean Héroard. Héroard was first recommended to the court in 1574, when Charles IX directed Ambroise Paré to select a physician who would dedicate himself to diseases of the horse. In addition to serving in this capacity under three kings, Héroard also continued to treat human patients. Some of his medical beliefs were unorthodox and he did not favor blood-letting. Héroard remained with Louis for 27 years and recorded every minute detail of his life.^{6,7} Henry IV was very close to his son and would play with him for hours. Marie de Medici, however,



The death of Louis XIII as depicted in an old engraving

showed little affection for her first-born and she never held or caressed the child. Louis XIII's upbringing was strict and he was often whipped for minor offenses. When Louis was eight and a half years old his father was assassinated by a religious fanatic. Louis XIII's childhood under his mother's regency appears to have been a nightmare. The queen mother and her intimate companion Concino Concini (who was married to her dwarf foster-sister) contrived to humiliate the boy. Concini had achieved enormous wealth and power under the Queen's sponsorship and showed his contempt for the young king by refusing to remove his hat in the youth's presence. He even went so far as to drain his wine cup at a royal banquet before the king raised his goblet.

Louis XIII's relationship with his wife, the infanta Anna of Austria, was also not very gratifying. The royal couple were both 14 when they

married. After the marriage was consummated, an act confirmed by the royal physician, Louis did not return to his bride for four years. Except for a relatively brief and apparently joyful period when he regularly shared his wife's company and bed, Louis' closest relationships were homosexual. In his later years he was infatuated with Henri d' Effiat, the Marquis de Cinq-Mars, a beautiful and vain youth who delighted in humiliating Louis XIII despite the titles and wealth bestowed upon him.

Louis XIII built the original residence at Versailles as a modest 15-room country retreat where he could escape from his royal responsibilities and compose and perform in the ballets that he loved. Yet he found little peace. His mother and his younger brother Gaston mounted half a dozen rebellions against the throne. Louis XIII was continually occupied in putting these down in addition to protecting his kingdom from English and Spanish invaders. Because there was no other successor to the throne, Gaston went unpunished. Guided by Cardinal Richelieu, Louis XIII did manage to cope with all of the threats to his reign, however, except his illness. Concini was killed while resisting arrest. The Marquis de Cinq-Mars was beheaded for his role in a plot, hatched with Gaston, to assassinate Louis, and for the seventh time Gaston had to beg his brother for forgiveness. France's borders were secured and the continuity of his reign was established in 1638 when the queen gave birth to a son.

It was the French custom not to give royal princes a name until their public baptism at the age of five. Confined to his bed, Louis XIII was unable to attend the public baptism of his dauphin. When the boy was brought to him after the ceremony the king was said to have asked his son, "What is your name now?" "Louis XIV, papa." "Not yet . . .," the king replied. Three weeks later he died.

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